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Case report

Severe Raynaud's phenomenon—A streamlined approach to acute management

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ABSTRACT

Raynaud's phenomenon is an exaggerated vasospastic response to cold or emotional stress which not only may cause the patient severe pain but also critical ischaemia and necrosis of the digits. We report the case of a 69-year-old woman who presented with rest pain, impending ulceration and necrosis of finger tips due to an episode of left-sided Raynaud's phenomenon. Intravenous prostacyclin was administered successfully as a bridge to endoscopic thoracic sympathectomy. Vascular surgery units are ideally positioned for the acute management of severe Raynaud's phenomenon to provide continuity of care to patients with profound digital ischaemia and impending tissue loss.

1. Introduction

Raynaud's phenomenon (RP) is an exaggerated vasospastic response to cold or to emotional stress that can be either primary (idiopathic) or secondary to a number of underlying conditions such as connective tissue disease, drugs or extrinsic vascular obstruction. The phenomenon is characterised by the sudden onset of cutaneous, acral vasospasm which results in cold extremities and an associated sharp demarcation in colour of the skin that exhibits a delay in restitution upon re-warming.

During an attack of RP the patient may experience intense pain, paraesthesia and loss of manual dexterity. If the vasospasm is particularly severe and long-lasting, the attack may be complicated by critical ischaemia and necrosis of the digits. Restoration of acral blood flow is therefore an emergency that requires co-ordinated and timely management. The following report demonstrates how vascular surgery is well placed as a specialty to manage Raynaud's crises acutely.

2. Report

In June 2009, A 72-year old, right hand dominant, female patient presented to the rheumatology clinic complaining of symptoms consistent with Raynaud's phenomenon (RP) involving all the fingers of her left hand over the past 10 years. On examination the fingers were blue at rest. Elevation of the hand produced pallor and on dependency, there was reactive hyperaemia of the digits. Trophic changes of the fingernails of the left side were evident and the left radial and ulnar pulses both palpable at the wrist.

Extensive investigation by the rheumatologists revealed no evidence of connective tissue disorder. Of the investigations for an embolic source or anatomical anomalies, magnetic resonance imaging revealed no evidence of a cervical rib or thoracic outlet syndrome; duplex ultrasonography showed no arterial or venous abnormality; and echocardiographic assessment demonstrated a structurally normal heart. Medical therapy in the form of topical nitrates, oral modified release nifedipine and oral sildenafil were initiated as first line therapy but either not tolerated or ineffective.

A month after the initial presentation, she presented to the emergency department with severe rest pain and impending ulceration of the distal phalax of the little finger from a severe left-sided Raynaud's phenomenon (Fig. 1). The patient was treated with a course of intravenous prostacyclin (Iloprost) over five consecutive days with significant improvement in her symptoms and no evidence of further tissue loss (Fig. 2). On discharge from the hospital, she was pain free and able to use her left hand for normal daily activities.

At the same time, the patient enquired for a definitive procedure for longer term symptomatic relief. The option of endoscopic thoracic sympathectomy (ETS) was discussed and agreed upon. The procedure was undertaken within a month of discharge from the hospital with excellent immediate effect. The patient remained pain-free with no further exacerbations of RP in subsequent follow-up visits.

3. Discussion

Raynaud's phenomenon represents a failure of acral vasomotor auto-regulation. To maintain adequate tissue perfusion, the bal-

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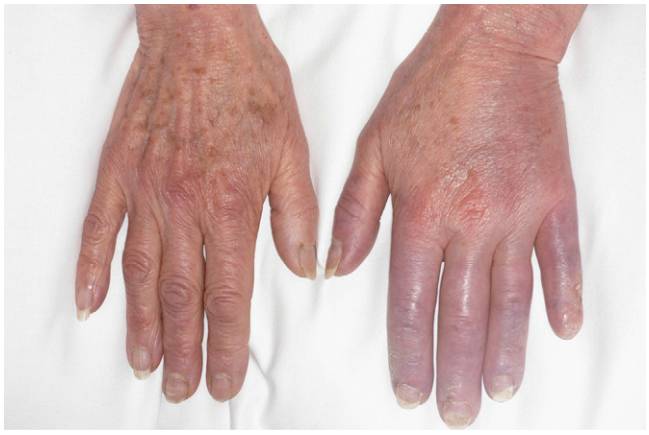

Fig. 1. Severe left sided Raynaud's phenomenon.

ance of vasodilatation and vasoconstriction is tightly regulated by local control mechanisms as well as neuro-hormonal mediators. Increased understanding of vascular biology and the pathophysiology of Raynaud's has lead to a number of different pharmacolgical and surgical approaches to management of the condition, including intravenous prostacyclin and ETS.

Prostaglandins are derivatives of arachidonic acid that possess many systemic actions, including vasodilatation and inhibition of platelet aggregation. Iloprost, synthetic prostaglandin I_2 (PGI_2), was first shown to be beneficial for the alleviation of Raynaud's phenomenon in the 1980s. Intravenous administration of iloprost, synthetic prostaglandin I_2 (PGI_2), is the first line therapy for patients with acute exacerbations of digital ischaemia and impending ulceration. A multi-centre, randomised, parallel placebo-controlled, double-blind study of a 6 h course of iloprost repeated daily for five consecutive days showed significant improvement in the condition including ulcer healing up to nine weeks post infusion.[1]

Surgical intervention for Raynaud's phenomenon is considered when medical therapy fails or is poorly tolerated, in this case with topical nitrates and modified release calcium channel blockers. One surgical option is ETS in which the sympathetic ganglia that supply the affected limb are divided in order to diminish the vasoconstrictive sympathetic activity in the limb and hence shift the balance of vascular tone away from constriction towards dilatation. Since the first large case series reported by Kux in 1951, ETS has been used effectively to treat several neurologic and vascular conditions, including Raynaud's phenomenon.[2]

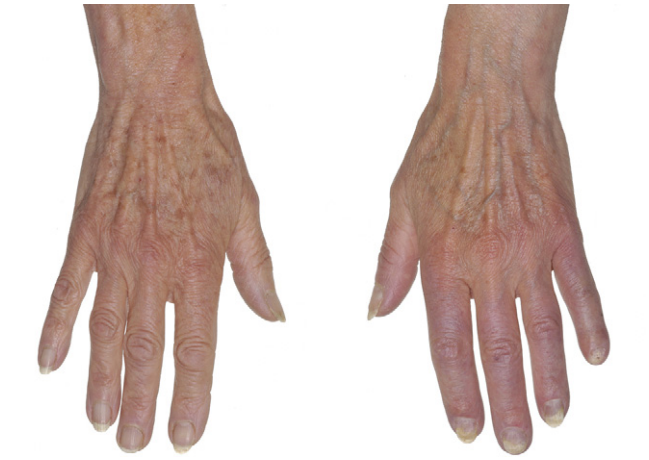

Fig. 2. Significant improvement after lioprost infusion.

ETS is associated with fewer side-effects such as permanent Horner's syndrome and neuralgia compared to an open approach and has gained acceptance as the preferred surgical technique for thoracodorsal sympathetomy. Though most case series reported immediate improvement in symptoms and healing of digital ulceration after ETS treatment for Raynaud's phenomenon, there remains controversy regarding the long-term efficacy of the procedure.[2] A recent longitudinal follow-up study of patients undergoing thoracoscopic sympathectomy found that one third of patients had a long-lasting benefit by 40 months of follow-up.[3]

Recurrence of symptoms after ETS treatment for Raynaud's phenomenon is likely to be multi-factorial and may not reflect failure of the procedure. Maga et al. examined the microcirculatory status in 25 patients who received ETS for Raynaud's disease during the 5 years post procedure. Basal capillary flow and maximal capillary refilling time were measured using laser-doppler flowmetry before the ETS procedure and at every visit during the 5-year follow up period. Significant improvements in the microcirculatory function to levels comparable to healthy volunteers were observed in these patients immediately after the ETS procedure and were maintained at every visit during follow up. However, when assessed using a subjective visual analogue scale, patients reported significant recurrence of symptoms as early as 6 months after the procedure despite recovery of normal microcirculatory function evident by the objective flowmetry assessments.[4]

In the setting of severe RP with impending tissue loss, ETS is uniquely placed to provide immediate term symptomatic relief and additional long term success in the prevention of morbidity related to digital ulceration. Matsumoto et al. reported a series of patients who undertook ETS as treatment for severe Raynaud's phenomenon refractory to medical therapy. There was significant symptomatic relieve in the initial post-operative period, although patients did report recurrence of symptoms in subsequent follow up. However, there was no recurrence of digital ulceration from this cohort of patients.[5]

4. Conclusion

This case highlights how a patient suffering from severe Raynaud's phenomenon can be managed successfully by a vascular surgery unit using iloprost infusion as a bridging therapy to endoscopic thoracic sympathectomy. Although multi-disciplinary evaluation of patient's with RP is essential, vascular surgeons are ideally placed to provide continuity of acute to long term care for patients with severe digital ischaemia and impending tissue loss.

Conflict of interest statement

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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